



Rhode Island Hospital
A Lifespan Partner

Department of Pathology

Lifespan Academic Medical Center
Rhode Island Hospital 593 Eddy Street Providence, RI 02903
Medical Director: [REDACTED]
M.D. [REDACTED]

Specimen: [REDACTED]
Procedure: 10/3/2003
Accessioned: 10/3/2003
Reported: 10/14/2003
Submitting Phys: [REDACTED]

Patient: [REDACTED]
Medical Record #: [REDACTED] Account #: [REDACTED]
DOB/Age/Sex: [REDACTED]
Location/Client: RIPRV / Rhode Island Hospital
Copy To: Wayland Square Surgicare
17 Seekonk Street
Providence, RI 02906

Additional Phys: [REDACTED]

Surgical Pathology Report

**** Procedures/Addenda Added ****

Final Diagnosis

- A. Skeletal muscle, left abductor longus, biopsy:
--Neurogenic atrophy, moderate (see comment).
- B. Skeletal muscle, left gluteus medius, biopsy:
--Neurogenic atrophy, mild to moderate (see comment).

Comments

(See microscopic description for complete details.) Taken together, the findings in both biopsies are most consistent with a neurogenic condition. The specimen in part A appears slightly more affected than that in part B; there are many more groups of obviously atrophic fibers in part A. The normal-staining patterns on immunohistochemical analysis of part A rules out the most common inherited muscular dystrophies.

Report Electronically Signed By [REDACTED] MD

Immunohistochemistry-AMC

Ordered: 10/14/2003

Reported: 10/14/2003

Paraffin block: A1

Staining for gamma-sarcoglycan also shows a normal-staining pattern.

Antibody	Result
Alpha-sarcoglycan (adhalin) (M; Ad1/20A6; Novocastra)	Normal-staining pattern
Beta-sarcoglycan (M; Bsarc/5B1; Novocastra)	Normal-staining pattern
Beta-dystroglycan (M; 43DAG1/8D5; Novocastra)	Normal-staining pattern
Dystrophin (C-terminus) (M; Dy2Dy8/6C5; Novocastra)	Normal-staining pattern
Dystrophin (rod domain) (M; Dy4/6D3; Novocastra)	Normal-staining pattern
Merosin laminin alpha 2 chain (M; Mer3/22B2; Novocastra)	Normal-staining pattern
Dysferlin (M; Ham1/7B6; Novocastra)	Normal-staining pattern
Dysferlin (M; Ham3/17B2; Novocastra)	Normal-staining pattern

Stains are performed using a standard streptavidin technique (Jackson) or a Vector ABC kit with a DAB chromogen. For monoclonal antibodies (denoted (M)) the specific clone utilized is indicated. The manufacturer of monoclonal (M) and polyclonal (P) primary antibodies is also included.

Rhode Island Hospital

Clinical History

27-year-old woman with bilateral, proximal, painless leg weakness. Genetic testing for spinal muscular atrophy is negative. Strength normal in quadriceps and hamstrings but has weak iliopsoas, gluteus medius, adductors, and gluteus maximus.

Pre-Operative Diagnosis

{Not Provided}

Post-Operative Diagnosis

{Not Provided}

Gross Description

- A) Received unfixed on ice in saline-moistened gauze labeled "Left Abductor Longus" is a fragment of soft, beefy-red tissue resembling skeletal muscle measuring approximately 2.0 x 1.5 x 0.3 cm in greatest dimensions. A 1-mm portion of the tissue is placed in glutaraldehyde for EM analysis (if necessary). Approximately half the tissue is snap-frozen in liquid nitrogen for histochemical analysis. The remainder of the tissue is formalin-fixed and submitted entirely in cassette A1. J-0
- B) Received unfixed on ice in saline-moistened gauze labeled "Left Gluteus Medius" is a fragment of soft, beefy-red tissue resembling skeletal muscle measuring approximately 2.0 x 1.7 x 0.3 cm in greatest dimensions. A 1-mm portion of the tissue is placed in glutaraldehyde for EM analysis (if necessary). Approximately half the tissue is snap-frozen in liquid nitrogen for histochemical analysis. The remainder of the tissue is formalin-fixed and submitted entirely in cassette A1. J-0

nlh/10/6/2003

Microscopic Description

A. H&E stain reveals moderate fiber-size variability, with grouped atrophy and scattered atrophic, hypertrophic, and angulated fibers. Within atrophic groups, nuclear knotting can be seen. There is no evidence of inflammation. A large amount of fat can be seen on the paraffin section. Trichrome stain fails to demonstrate classic ragged-red fibers; atrophic fibers are staining red, however. Scattered subsarcolemmal staining of mitochondria can be seen, which is shown more prominently on NADH and SDH stains. Those stains also reveal many moth-eaten fibers, but no target fibers. ATPase stains reveal fiber-type grouping of both fiber types. PAS stain reveals normal amounts of glycogen that is appropriately digested with diastase. Oil Red O stain demonstrates normal amounts of lipid. Myophosphorylase stain is positive. Immunohistochemical analysis reveals normal-staining patterns for dystrophins 1 and 2; alpha-, beta-, and gamma-sarcoglycans; beta-dystroglycan; merosin; and dysferlin 1 and 2.

B. H&E stain reveals mild to moderate fiber-size variability, with scattered atrophic, hypertrophic, and angulated fibers. Rare groups of fibers can be seen that are mildly atrophic, though this grouped atrophy is not as obvious as it is in part A. There is no evidence of inflammation. A large amount of fat can be seen on the paraffin section. Trichrome stain fails to demonstrate classic ragged-red fibers. Scattered subsarcolemmal staining of mitochondria can be seen, which is shown more prominently on NADH and SDH stains. Those stains also reveal many moth-eaten fibers, but no target fibers. ATPase stains reveal a marked type I fiber predominance with rare, focal grouping of type II fibers. PAS stain reveals normal amounts of glycogen that is appropriately digested with diastase. Oil Red O stain demonstrates normal amounts of lipid. Myophosphorylase stain is positive.